Thoracic aneurysm endovascular repair of extremely tortuous aorta in neonatal Marfan syndrome patient with major scoliosis

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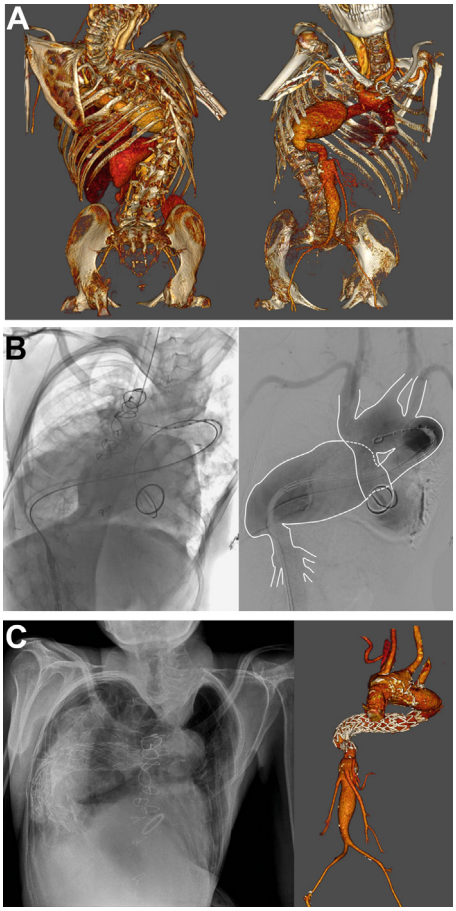
A 27-old man was referred to our Marfan clinic for adult follow-up. He had been followed up in a pediatric center for neonatal Marfan syndrome (MFS) with an *FBN1* gene mutation (premature termination codon variant). His history included ascending aortic repair by the Yacoub procedure at 5 years old, followed by a Bentall procedure with mechanical aortic valve and mechanical mitral valve replacement at the age of 10 years. He also underwent distal aortic arch open repair and type IV thoracoabdominal open repair at 17 and 19 years old, both for aneurysms.



At the current presentation, he had a major spinal deformity with severe restrictive lung disease (*A*/Cover, *Left Panel*). Follow-up computed tomography angiography highlighted a 64-mm nondissecting aneurysm of the descending aorta below the arch graft and above the thoracoabdominal graft (*A*/Cover, *Right Panel*). After multidisciplinary assessment, the patient was considered unfit for open repair owing to his restrictive respiratory insufficiency, and thoracic endovascular repair (TEVAR) was decided in accordance with the guidelines.[1] The use of TEVAR for MFS patients remains controversial when endografts are landed in the native aorta. The risk of landing zone complications can be reduced using a dedicated protocol for stent graft sizing, including minimal proximal oversizing, the use of stent grafts without barbs and bare stents, and the graft-in-graft technique whenever possible.[2] Complex aortic angulation is best exposed, not via a left oblique anterior view but via an anteroposterior view with the C-arm (*B, Left Panel*). The patient provided written informed consent for the report of his case details and imaging studies.

Two tapered stent grafts (31-25 × 207 mm; Valiant Captivia; Medtronic) were delivered through an extra-stiff guide wire (Lunderquist wire; Cook Medical Inc) and after crossing both major angulations (*B, Right Panel*). The through-and-through technique was not needed but was anticipated in case it had been needed. A postoperative chest radiograph showed both stent grafts in the chest (*C, Right Panel*). His postoperative course was marked by a bilevel positive airway pressure, noninvasive ventilation requirement for 3 days. The 2-year postoperative computed tomography angiogram confirmed aneurysm exclusion without complications at the landing zones (*C, Left Panel*).

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Neonatal MFS represents the most severe end of the phenotypic spectrum of the syndrome.[3] The incidence of nondissecting aortic, visceral, or peripheral aneurysms in those with MFS is unknown and probably underestimated. Almost 25% of MFS patients present with aortic branch aneurysms, as reported in a larger study.[4] Nondissecting aneurysms are highly associated with a more severe cardiovascular phenotype with a significantly greater risk of aortic events (aortic surgery and acute aortic dissection).[4,5] Aortic tortuosity increases in proportion to the degree of scoliosis[6] and is associated with a more severe MPS phenotype.[7] It can result in technical pitfalls for surgeons such as delivery system progression difficulties and a lack of apposition of the stent graft during TEVAR.

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